

Bilateral choanal atresia in a newborn infant

Obostrana atrezija hoana u novorođenčeta

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Abstract. Aim: Bilateral nasal obstruction in neonates is a potentially fatal condition since neonates are obligatory nasal breathers. Congenital choanal atresia is a result of the persistence of the embryological bucconasal membrane, which separates the nasal cavity and the nasopharynx in the early embryological development. The aim of this article is to present this rare anomaly which can be lethal in neonatal period. **Case report:** A female neonate was born at General Hospital Pula and soon after birth signs of respiratory distress and intermittent cyanosis could be seen. Since clinical condition and oxygen saturation on room air improved when crying, the bilateral choanal atresia was suspected. Oropharyngeal airway was inserted to make the airway patent. Insertion of a feeding tube via the nostrils was not possible. Otorinolaringologist was consulted and confirmed the diagnosis. The infant was transferred to a tertiary pediatric center where CT scan of the choanal region was performed, showing membranous bilateral choanal atresia. The transnasal endoscopic operation was performed and stents were put in the nostrils. Stents were removed after 11 days. The child was released home in good condition without any signs of respiratory compromise in the follow up period of 2 years. **Conclusions:** Bilateral choanal atresia is a neonatal emergency. The anomaly can be surgically corrected.

Key words: choanal atresia; endoscopy; neonate; transnasal approach

Sažetak. Cilj: Novorođena djeca obligatorni su nosni disači. Stoga obostrana opstrukcija nosnih hodnika u novorođenačkoj dobi može biti po život opasno stanje. Prikazat ćemo slučaj novorođenčeta s obostranom atrezijom hoana. Cilj ovog članka je prikazati anomaliju koja je izuzetno rijetka u novorođenačkoj dobi, a koja može dovesti do letalnog ishoda. **Prikaz slučaja:** Terminko žensko novorođenče rođeno je u Općoj bolnici Pula i odmah po porodu bili su prisutni znakovi respiratornog distresa. Kako su se kliničko stanje i saturacija kisikom poboljšavali kada bi novorođenče plakalo, postavljena je sumnja na obostranu atreziju hoana. Postavljen je orofaringealni „airway“ kako bi se olakšalo disanje djeteta. Nazogastričnu sondu nije bilo moguće postaviti zbog otpora u nosnim hodnicima. Konzultiran je otorinolaringolog koji je potvrdio dijagnozu. Novorođenče je premješteno u Tercijarni pedijatrijski centar (KBC Rijeka) gdje je učinjen CT hoanalne regije na kojem se vidi obostrana membranozna atrezija hoana. Učinjen je operativni zahvat transnazalnom endoskopskom tehnikom i u svaku nosnicu je postavljen stent. Stentovi su uklonjeni jedanaestog postoperativnog dana. Novorođenče je otpušteno kući urednog kliničkog statusa. U daljnje dvije godine praćenja nije bilo znakova respiratornih tegoba. **Zaključak:** Obostrana atrezija hoana je hitno stanje koje u novorođenačkoj dobi može imati i letalni ishod. Anomalija se liječi kirurški.

Ključne riječi: atrezija hoana; endoskopija; novorođenče; transnazalni pristup

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INTRODUCTION

Bilateral nasal obstruction in neonates is a potentially fatal condition since neonates are obligatory nasal breathers. Congenital choanal atresia is a result of the persistence of the embryological bucconasal membrane, which separates the nasal cavity and the nasopharynx in the early embryological development. The incidence ranges between 1 in 5000 to 10000 live births. Choanal atresia is bilateral in 45% of cases, and of the

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unilateral cases, 71% involves the right choanae. It can be bony, membranous or mixed. Male to female ratio is approximately 2:1¹⁻⁴. Emergency treatment consists of placing a plastic airway into the mouth to keep it open⁵. Many surgical methods are available for the correction of choanal atresia (transpalatal, transseptal and transnasal)^{2,6}.

CASE REPORT

We present a case report of a female infant born with bilateral choanal atresia at General hospital Pula. It was the first mother's pregnancy; during the pregnancy, mother did not use any drugs, didn't smoke or take any alcohol, was not exposed to X-rays.

The birth was performed using Kiwi complete vacuum delivery system at 41 weeks of gestation. The infant weight was 3640 g, length 50 cm and head circumference 34 cm. Apgar score was 6/8/9. Respiratory distress and intermittent cyanosis were present soon after birth. Since clinical condition and oxygen saturation on room air improved when crying, the bilateral choanal atresia was suspected. Oropharyngeal airway was inserted to make the airway patent. Since insertion of a feeding tube via the nostrils was not possible, otorhinolaryngologist was consulted and confirmed the diagnosis.

The infant was transferred to a tertiary care pediatric center (University Hospital Center Rijeka, Department of pediatrics, Pediatric Intensive Care Unit (PICU)) for a further diagnostic and therapeutic management. Oropharyngeal airway was used to make the airway patent during the transport.

After arrival to PICU the otorhinolaryngologist was consulted and computed tomography scan of the choanal region was performed, showing membranous bilateral choanal atresia (figure 1). The endoscopic operation was performed and stents were put in the nostrils. Stents were removed after 11 days. Two days later the child was released home in good condition without any signs of respiratory compromise in the follow up period of two years.



Figure 1. Axial CT images demonstrate thin membranes (arrows) occluding the choanal lumen. No ossification is seen. Fluid is seen above the obstruction point.

DISCUSSION

Choanal atresia can be the only anomaly found in patients or can be associated with other anomalies. Five percent of patients have monogenic syndromes or conditions. For example, the CHARGE syndrome consists of Coloboma of the eye, Heart defects, Atresia of the choanae, Retardation of growth and development, Genital or urinary abnormalities, Ear abnormalities or deafness⁷. Using data published data from the National Birth Defects Prevention Study, Kancherla et al

(2014) found evidence linking choanal atresia to maternal exposure to various nutrients, thyroid medications and cigarettes⁸.

The differential diagnosis includes deviated nasal septum, dislocated nasal septum, septal hematoma, mucosal swelling, encephalocele, nasal dermoid, hamartoma, cordoma and teratoma⁹.

Unilateral cases of choanal atresia are often diagnosed later in life; the patients complain of unilateral nasal obstruction, often following an upper respiratory tract infection. Bilateral congenital choanal atresia is a neonatal emergency because neonates are obligatory nasal breathers. The suspicion of bilateral choanal atresia must arise in every neonate presenting with signs of respiratory distress and intermittent cyanosis improving when crying. The airway is kept patent by an oral airway. The soft rubber nipple can be used; the tip of the nipple is cut and the nipple is put in the neonate's mouth^{1,2,5}.

The diagnose can be made clinically (respiratory distress, intermittent cyanosis, better clinical condition when the neonate is crying, failure to pass a feeding tube through both nostrils) but for a confirmation of the diagnosis nasal endoscopy and a CT scan of the choanal region must be performed.

The surgery must be performed in the first weeks of life. Various approaches have been described (transnasal, transseptal, transpalatal) but nowadays the preferred approach is the transnasal endoscopic route^{2,6}. The duration of stenting remains controversial. Most authors advise stenting for 3-8 weeks^{2,3,5}. Abbelee et al. in 2002 showed that good results can be achieved even with a very short stenting time. They performed transnasal endoscopic surgery in 40 children aging 3 days to 15 years. The stents were put in the nostrills and removed two days after surgery in all cases. In 80% of cases the nasal patency was normal during the follow up period⁶. In our case, the stents were removed after 11 days and the child was released home two days later and had no signs of respiratory compromise during the follow up period of two years.

Although, according to the literature, the incidence of choanal atresia in the neonatal period is 1:5000-10000, in General Hospital Pula (Pula, Croatia) there were no neonates born with choanal atresia in the last 40 years. The number of births in the above mentioned period was 60 – 70 thousands. We can conclude that the above mentioned anomaly in our region is extremely rare but it is necessary to think about it in a neonatal period since it can lead to a lethal outcome.

CONCLUSIONS

Bilateral choanal atresia is a neonatal emergency since neonates are obligatory nasal breathers. The anomaly can be successfully surgically corrected by transnasal, transseptal or by transpalatal approach. Transnasal endoscopic route is the preferred surgical approach nowadays.

Conflicts of interest statement: The authors report no conflicts of interest.

REFERENCES

1. Asma A, Roslenda AR, Suraya A, Saraiza AB, Aini AA. Management of congenital choanal atresia (CCA) after multiple failures: A Case Report. *Med J Malaysia* 2013; 68:76-8.
2. Cumberworth VL, Djazaeri B, Mackay IS. Endoscopic fenestration of choanal atresia. *J Laryngol Otol* 1996;109: 31-5.
3. Shiva Kumar AM, Naik AS, Praveen DS. Choanal atresia: experience with transnasal endoscopic technique. *Indian J Otolaryngol Head Neck Surg* 2005;57:96-8.
4. Leclerc JE, Fearon B. Choanal atresia and associated anomalies. *Int J Pediatr Otorhinolaryngol* 1987;13: 265-72.
5. Saleem AF, Ariff S, Aslam N, Ikram M. Congenital bilateral choanal atresia. *J Pak Med Assoc* 2010;60:869-72.
6. Van Den Abbelee T, Francois M, Narcy P. Transnasal endoscopic treatment of choanal atresia without prolonged stenting. *Arch Otolaryngol Head Neck Surg* 2002; 128:936-40.
7. Trider CL, Blake K. Obstructive Sleep Apnea in a Patient with CHARGE Syndrome. *Case Rep Otolaryngol online* 2012;2012:907032.
8. Kancherla V, Romitti PA, Sun L, Carey JC, Burns TL, Siega-Riz AM et al. Descriptive and risk factor analysis for choanal atresia: The National Birth Defects Prevention Study, 1997-2007. *Eur J Med Genet* 2014;57:220-9.
9. Gnagi SH, Schraff SA. Nasal obstruction in newborns. *Pediatr Clin North Am* 2013;60:903-22.